SURVEILLANCE OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS:

RECOMMENDATIONS FROM THE BRAIN TUMOR WORKING GROUP

September 1998

Brain Tumor Working Group

National Coordinating Council for Cancer Surveillance

This report was prepared by the Brain Tumor Working Group of the National Coordinating Council for Cancer Surveillance, and edited by Robert R. German, M.P.H.; Brooke Steele, D.O.; and Gayle Clutter, R.T., C.T.R. The Brain Tumor Working Group was comprised of the following people listed alphabetically:

Sally A. Bushhouse, D.V.M., Ph.D.
Director
Minnesota Cancer Surveillance System

Rosemarie Clive, L.P.N., C.T.R.
Cancer Department Director
American College of Surgeons

Gayle Clutter, R.T., C.T.R.
Former President
National Cancer Registrars Association
Program Analyst
Centers for Disease Control and Prevention

Faith Davis, Ph.D.
Professor and Director
Div. Epi./ Biostat. Sch. Public Health
University of Illinois at Chicago
Director
Central Brain Tumor Registry of the United States

Robert R. German, M.P.H.
Epidemiologist
National Program of Cancer Registries
Centers for Disease Control and Prevention

Susan Gershman, Ph.D.
Director
Massachusetts Cancer Registry

James G. Gurney, Ph.D.
Department of Pediatrics
Division of Epidemiology/Clinical Research
University of Minnesota

Holly L. Howe, Ph.D.
President
North American Association of Central Cancer Registries
Director
Illinois State Cancer Registry

Sarah H. Landis, M.P.H.
Epidemiologist
American Cancer Society

Edward R. Laws, Jr., M.D., F.A.C.S.
Professor of Neurosurgery
Professor of Medicine
University of Virginia, Charlottesville

Herman R. Menck, M.B.A.
Clinical Information Manager
National Cancer Data Base
American College of Surgeons

Constance Percy, M.S.P.H.
Statistician
Surveillance, Epidemiology, and End Results Program
National Cancer Institute

Jerri Linn Phillips, M.A., C.T.R.
Research Analyst
National Cancer Data Base
American College of Surgeons

Lynn Ries, M.S.
Health Statistician
Surveillance, Epidemiology, and End Results Program
National Cancer Institute

Brooke Steele, D.O.
Medical Epidemiologist
National Program of Cancer Registries
Centers for Disease Control and Prevention

Tanya S. Surawicz, M.P.H.
Research Associate
Central Brain Tumor Registry of the United States

Preface

This report on Surveillance of Primary Intracranial and Central Nervous System Tumors was prepared for the National Coordinating Council for Cancer Surveillance (NCCCS) by an appointed Working Group of experts in brain tumor epidemiology and in cancer registration, and representatives of several public and private organizations involved in cancer surveillance in the United States. The Working Group described the clinical and epidemiologic significance of benign and malignant intracranial and central nervous system (CNS) tumors; reviewed the current status of nonmalignant brain tumor surveillance among cancer registries in the United States; analyzed data for brain and other CNS tumors, as well as non-CNS intracranial tumors; and, assessed the feasibility of routine collection of data for nonmalignant brain tumors.

Based on its review, analysis, and assessment, the Working Group prepared a report with recommendations for the NCCCS regarding data collection of all primary intracranial and CNS tumors (see Executive Summary). The NCCCS discussed the report and recommendations at its September 9, 1998 meeting. The Council voted to accept two recommendations of the Working Group: (1) to derive a standard definition for all primary intracranial and CNS tumors and (2) to develop a standard site and histology definition for tabulating and estimating rates of these tumors. The NCCCS deferred further consideration and action on the two remaining Working Group recommendations.

National Coordinating Council for Cancer Surveillance

September 1998

TABLE OF CONTENTS

EXECUTIVE SUMMARY

The National Coordinating Council for Cancer Surveillance established the Brain Tumor Working Group (BTWG) to examine current reporting practices for brain tumors among cancer registries in the United States. This anatomic site was selected for a special review because much morbidity and mortality is associated with both malignant and nonmalignant brain tumors. The BTWG determined that the review should encompass the brain and other parts of the central nervous system (CNS), as well as non-CNS intracranial tumors. Four reporting sources -- the Central Brain Tumor Registry of the United States (CBTRUS), the Minnesota Cancer Surveillance System (MCSS), the National Cancer Data Base (NCDB), and the Surveillance, Epidemiology, and End Results (SEER) Program -- contributed data to the BTWG for analysis. The major findings are highlighted in this summary, and recommendations regarding the collection of data for primary intracranial and CNS tumors are provided.

More than 28,000 new cases of primary malignant and benign brain tumors were diagnosed nationwide in 1995.[1] Approximately 12,000 people died of invasive brain tumors and 947 died of benign brain tumors during that year.[2] There were also 131 deaths due to tumors of uncertain behavior and 2,788 deaths due to tumors of unspecified behavior reported for those sites. From 1979-1995 mortality rates have remained relatively stable for invasive, benign, and unspecified tumors (Fig. 1). The NCDB, which reports hospital registry data, recently reported 5-year survival rates for patients diagnosed with brain tumors from 1985-1988 and from 1990-1992.[3] Five years after diagnosis, approximately 22 percent of patients with malignant tumors and 72% of patients with nonmalignant tumors were alive. The clinical outcome of both malignant and nonmalignant tumors, however, may also depend on factors unrelated to behavior.

For example, survival rates are generally higher for benign meningiomas than for malignant meningiomas (Fig. 2), but treatment of meningiomas may be limited by their location. Favorably situated lesions (e.g., lateral sphenoid wing) are usually amenable to complete removal, whereas basal meningiomas are more difficult to fully and safely excise.[4]

Only 15 state registries (Appendix A) collected data for benign intracranial and CNS tumors in 1997. Three SEER areas also collect information about these tumors, but they do not currently report it. Comparison of the data by cancer registries, however, is made difficult by the lack of standard site groupings and histology groupings for the coding systems (i.e., International Classification of Diseases for Oncology, World Health Organization) that most of them use.

Nonmalignant tumors constituted a significant percentage of the primary intracranial and CNS tumors reported by the data sources. For CBTRUS, from 1990-1993 approximately 46 percent of the tumors reported for these sites were nonmalignant (Fig. 3). Fifty-one percent of the primary intracranial and CNS tumors reported by MCSS from 1989-1994 were nonmalignant, and NCDB reported more than 33 percent as nonmalignant during that period.

Histologically, much variation occurs among nonmalignant primary intracranial and CNS tumors. While the majority of malignant tumors reported by all of the sources were of neuroepithelial origin (Table 1), most of the nonmalignant tumors were in the meninges.

For registries that consider making nonmalignant intracranial and CNS tumors reportable, a twofold increase in the overall number of CNS cases could be expected. In addition to the increased costs associated with the increased workload, registries should also be aware that additional training of registrars, new registry manuals, modifications of case finding methods, modifications in the registries' database and data processing software, and revisions in legislation

and regulations would be needed. A mandate to report would be required to ensure high quality surveillance.

Based on its findings the BTWG has prepared four recommendations regarding data collection for primary intracranial and CNS tumors:

1. **We recommend** that the following standard definition be used for collecting precise data for all primary intracranial and CNS tumors:

   Primary intracranial and CNS tumors are all primary tumors occurring in the following sites, irrespective of histologic type or behavior: brain, meninges, spinal cord, cauda equina, cranial nerves and other parts of the CNS, pituitary gland, pineal gland, and craniopharyngeal duct (see Appendix B).

2. **We recommend** the development of a standard site and histology definition for tabulating estimates of these tumors to allow comparability of information across registries. Pathologists, the North American Association of Central Cancer Registries (NAACCR), the Commission on Cancer (COC), the Surveillance, Epidemiology, and End Results (SEER) Program, the National Program of Cancer Registries (NPCR), and the International Agency for Research on Cancer (IARC) need to be involved in developing this standard.

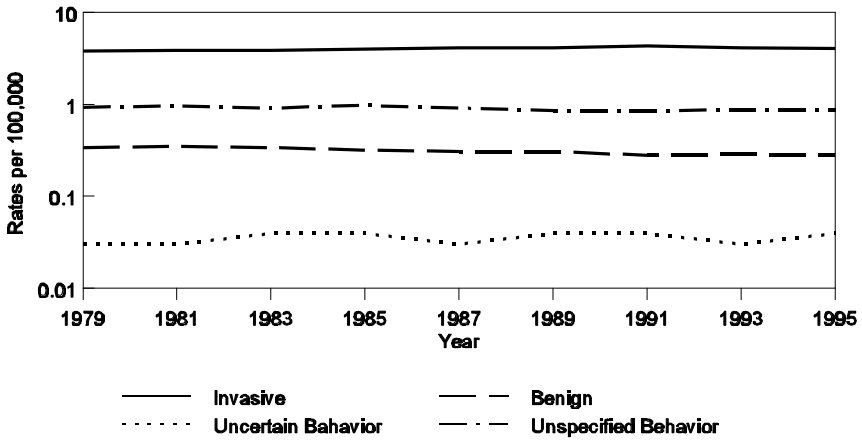
3. **We recommend** collection of data for primary intracranial and extracranial CNS tumors by all registries, hospital- and population-based. This effort will necessitate a change in the COC requirements and will increase costs to the hospital-based programs. Federal funding should be allocated to supplement the additional transition and ongoing data collection costs that will be incurred by central registries. Before

additional data collection is implemented, a pilot study should be conducted in multiple states to assess the procedures and quality control functions needed, as well as the costs of collecting data on these tumors.

4. **We recommend** that the appropriate government and professional organizations be involved in carrying out the development and implementation of special training programs and curricula for central registry, hospital registry, and laboratory personnel, as well as the development of computerized edit-checking procedures. Training for reporting and tabulating primary intracranial and CNS tumors should be offered on a regular basis.

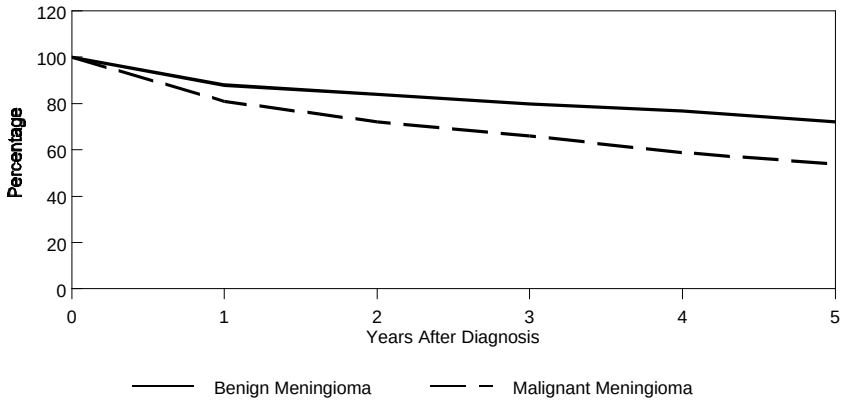
EXECUTIVE SUMMARY: FIGURES AND TABLE

Figure 1. Brain and Other Nervous System Mortality Rates by Behavior, National Center for Health Statistics, 1979-1995



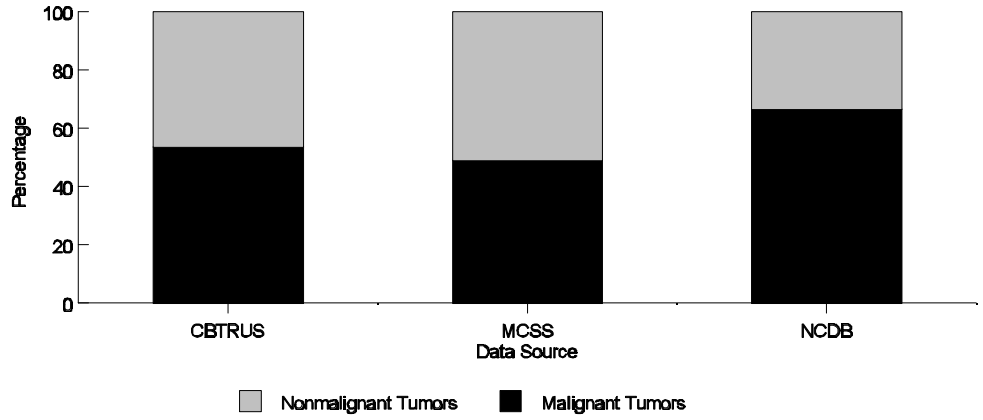
Note: Rates are age-adjusted to the 1970 U.S. standard

Figure 2. Percentages of Cases Surviving at One, Two, Three, Four, and Five Years After Diagnosis, National Cancer Data Base (NCDB), 1985-1990



EXECUTIVE SUMMARY: FIGURES AND TABLE (continued)

Figure 3. Percentages of Malignant and of Nonmalignant (Benign, Uncertain Behavior) Primary Intracranial and Central Nervous System Tumors, Central Brain Tumor Registry of the United States (CBTRUS), Minnesota Cancer Surveillance System (MCSS), and the National Cancer Data Base (NCDB)



Note: Reported data are incident cases from 1989-1994 except for CBTRUS, which is 1990-1993**.** MCSS cases are limited to microscopically-confirmed, and NCDB cases are from hospitals that report nonmalignant cases**.**

Table 1. Numbers and Percentages of Histologic Grouping by Malignant and by Nonmalignant Primary Intracranial and Central Nervous System Tumors, Central Brain Tumor Registry of the United States (CBTRUS), Minnesota Cancer Surveillance System (MCSS), and the National Cancer Data Base (NCDB)

Histologic Grouping	CBTRUS		MCSS		NCDB	
	Malignant	Nonmalignant	Malignant	Nonmalignant	Malignant	Nonmalignant
	Number %	Number %	Number %	Number %	Number %	Number %
Neuroepithelial	6,477 (87.4)	275 (4.3)	1,689 (89.6)	91 (4.6)	39,701 (85.8)	1,064 (4.6)
Cranial/Spinal Nerves	12 (0.2)	980 (15.3)	3 (0.2)	397 (20.2)	108 (0.2)	2,618 (11.3)
Meninges	137 (1.8)	3,509 (54.8)	25 (1.3)	941 (48.0)	994 (2.1)	13,667 (59.0)
Sellar region	20 (0.3)	1,444 (22.6)	11 (0.6)	510 (26.0)	100 (0.2)	4,749 (20.5)
Other	767 (10.3)	193 (3.0)	156 (8.3)	22 (1.2)	5,381 (11.7)	1,077 (4.6)
TOTAL	7,413 (100)	6,401 (100)	1,884 (100)	1,961 (100)	46,284 (100)	23,175 (100)

Note: Incidence data are from 1989-1994 except for CBTRUS cases, which are from 1990-1993. MCSS cases are limited to microscopically confirmed, and NCDB's cases are from hospitals that report nonmalignant cases.

REFERENCES

1. CBTRUS (1996) First annual report 1995. Central Brain Tumor Registry of the United States. Chicago, Illinois.
2. National Center for Health Statistics, public use mortality files for 1979-1995.
3. Surawicz TS, Davis F, et al. Brain tumor survival: results from the National Cancer Data Base. (Accepted for publication in Journal of Neuro-Oncology).
4. Thapar K, Laws ER. Tumors of the central nervous system. In Murphy GP, Lawrence W Jr, Lenhard RE Jr, eds. Textbook of clinical oncology, ed 2. Atlanta, American Cancer Society 1995;378-410.

INTRODUCTION

Because nonmalignant brain tumors often can have a malignant clinical course leading to substantial long-term morbidity and high risk for death, the distinction between histopathologically-defined malignancies versus nonmalignancies is tenuous from both a clinical and an epidemiologic standpoint. Likewise, because nonmalignancies have not been systematically or consistently included in most population-based cancer surveillance systems, the public health community has little information to offer on either incidence or mortality patterns and trends of nonmalignant brain tumors in the United States (U.S.). Additionally, there have been virtually no analytic epidemiology research studies conducted that would help identify factors that influence the risk for nonmalignant brain tumor occurrence. Thus, despite their clinical importance, both the public health surveillance mission and the etiologic research mission of cancer prevention and control are compromised by the exclusion of nonmalignant brain tumors in registry systems. Recognizing this challenge, the National Coordinating Council for Cancer Surveillance (NCCCS) established a Brain Tumor Working Group (BTWG) during its semi-annual meeting in January 1997. The purpose of BTWG was to review the status of the surveillance of brain tumors in the U.S. and examine the feasibility of collecting data for nonmalignant brain tumors. Upon completion of the project, recommendations regarding surveillance of both malignant and nonmalignant (i.e., benign or uncertain behavior) brain tumors would be presented at a special meeting of the NCCCS. This report describes data collection by U.S. registries for primary intracranial and central nervous system (CNS) tumors. It includes data from several registries to demonstrate variability in current reporting practices.

BACKGROUND

Classification

Historically, cancer registries have used the International Classification of Diseases (ICD) system, which describes tumors by their location (topography) and behavior (benign versus malignant),[1] to code neoplasms. Tumor nomenclature describing histology was developed by pathologists and incorporated into the International Classification of Diseases for Oncology (ICD-O) system as a morphology code in 1976[2] and revised in a second edition (ICD-O-2).[3] However,

not all of the morphology codes in the current ICD-O classification used by cancer registries are consistent with the recent World Health Organization (WHO) brain tumor codes.[3,4] The recent WHO brain tumor classification has new terms that are not included in ICD-O-2; however, a new third edition to ICD-O may include these terms. ICD-O-2 also includes terms which are not categorized by WHO.

The CNS environment also contributes to the difficulty in characterizing tumors that involve this site. In contrast to tumors arising in other organ systems, the terms "benign" and "malignant" are only relative distinctions for CNS neoplasms. Depending on location, histologically benign CNS tumors can result in similar or worse outcomes compared with malignant tumors. For these and other reasons, classic oncologic concepts predicated on histologic grade, nodal status, and staging strategies are not entirely applicable to tumors of the CNS.[5]

When the BTWG discussed whether to consider anatomic sites other than the brain for this report, the unique features of the CNS and the morbidity and mortality associated with non-CNS intracranial tumors were closely examined. For the sake of simplicity in reporting and analyzing data for all of these sites, the designation of "primary intracranial and CNS tumors" was suggested. This designation, proposed by Walker et al,[6] is used by the Central Brain Tumor Registry of the United States (CBTRUS) and includes the brain, cerebral meninges, cranial nerves, pituitary gland, craniopharyngeal duct, pineal gland, spinal cord, cauda equina, and spinal meninges. The intracranial sites are the brain, cerebral meninges, cranial nerves and other intracranial parts of the CNS; the craniopharyngeal duct; and the pineal and pituitary glands. The extracranial sites are the spinal cord, cauda equina, and spinal meninges. The BTWG also included lymphomas in the histology groupings for these sites. The topography codes for the sites are in Appendix B, and the histology groupings are listed in Appendix C. In this report, no distinctions are made between different age groups for intracranial and CNS tumors.

Incidence

The age-adjusted incidence rate for malignant brain and other CNS tumors in the U.S. for 1991-1995 was 6.1 per 100,000 person years.[7] Approximately 17,400 new cases will be diagnosed in 1998.[8] The incidence of nonmalignant brain and other CNS tumors is difficult to

ascertain because few cancer registries collect or report these data. However, CBTRUS reported an incidence rate for all primary brain and CNS tumors, including the pituitary and pineal glands, of 11.8 per 100,000 for 1990-1993;[9] the organization also estimated that 28,600 new cases of primary malignant and benign brain tumors were diagnosed nationwide in 1995.[10] For both men and women, rates for malignant and nonmalignant brain tumors decline after a peak in childhood (younger than 10 years), increase after age 25, and stabilize after age 75; overall rates are higher for males.[11]

Although the Surveillance, Epidemiology, and End Results (SEER) Program reported a 2.8 percent decrease in the incidence of invasive brain and other nervous system cancer from 1991 to1995,[7] within the last decade there have been many reports of dramatic increases in the incidence of brain tumors,[12-21] particularly among children[22,23] and the elderly.[12,16,17] Some consider the increased rates as histology-specific[14] and indicative of a true shift in incidence.[13] Other studies have concluded that many of the new cases are an artifact of changing diagnostic procedures.[16,17] In the U.S. the discussion regarding the increased incidence of brain tumors has been limited to rates for malignant tumors. Standard reporting practices would aid in the interpretation of time trends.

Molecular studies[24-26] have demonstrated that some low grade or benign intracranial tumor subtypes transform to malignant tumors. To understand the factors that might contribute to this transformation and whether incidence rates for both malignant and nonmalignant intracranial tumors are affected, the full spectrum of the disease needs to be observed.

Mortality

Brain tumors, regardless of behavior, are the second leading cause of death from neurological disease.[27] In 1995, malignant brain tumors accounted for 12,062 deaths, and 947 deaths were due to benign brain tumors. Another 131 deaths were reported for brain tumors of uncertain behavior, and 2,788 persons died of brain tumors for which the behavior was unspecified.[28] The estimated number of deaths in 1998 due to malignant brain and other CNS tumors is 13,300.[8] Increased mortality rates for malignant brain tumors, particularly among the elderly, have been reported. One study concluded that the increases were largely

related to better diagnostic technology and the introduction of support programs such as Medicare that facilitate diagnostic procedures in the elderly.[17] Another study attributed the rising primary malignant brain tumor mortality to differential survival and its effect on the surviving gene pool in an aging population.[28]

Survival

Prognosis for CNS tumors depends on at least four variables: tumor histopathology, anatomic location, patient age, and neurologic status.[5] According to a recent analysis using SEER data, from 1973-1991 overall survival rates for malignant brain tumors and rates for patients with three specific histological types -- astrocytoma, medulloblastoma, and oligodendroglioma -- improved.[29] Few data are available for survival rates for nonmalignant primary intracranial and CNS tumors. However, Surawicz et al used data from the National Cancer Data Base (NCDB), a joint project of the Commission on Cancer (COC) of the American College of Surgeons (ACoS) and the American Cancer Society, to examine survival rates for patients diagnosed from 1985-1988 and from 1990-1992 with malignant or benign brain tumors;[30] NCDB collects data from hospital tumor registries. Based on the records of more than 60,000 patients, the authors found a 21.6 percent 5-year survival rate for patients with malignant tumors and a 72.4 percent rate for those with benign tumors. The most favorable prognosis was associated with neurilemmomas, pilocytic astrocytomas, and meningiomas. In contrast, microgliomas, lymphomas, malignant gliomas, and anaplastic astrocytomas were associated with a poor outcome. Surawicz et al also found variations in survival for some tumors depending on location. For example, survival rates for glioblastomas, which generally have a poor prognosis, improved if the tumors were located in the cerebellum; similarly, astrocytomas and anaplastic astrocytomas were associated with a better outcome if the tumors were in the ventricles or the cerebellum.

Risk Factors

Although malignant and nonmalignant intracranial and CNS tumors have undergone considerable study, etiologic and pathophysiologic details concerning their genesis remain obscure. Definite genetic predispositions for the development of brain tumors have been identified; however, population-based studies suggest that no more than 4 percent of these

tumors can be attributed to heredity.[31] Several environmental carcinogens may be associated with brain tumors, including ionizing radiation,[11] electromagnetic fields,[32] and pesticides;[33] sustained exposure to vinyl chlorides, polycyclic hydrocarbons, and nitrosoureas has been implicated as well.[5] Also, the presence of the Epstein-Barr virus contained in the DNA of primary lymphoma suggests that a viral etiology for human brain tumors cannot be entirely ignored.[5] Accurate and complete data are necessary to develop hypotheses to identify the causes of intracranial and CNS tumors. The heterogeneity of brain tumors may mask the identification of causes when histology-specific studies are limited by the number of available cases.

Surveillance

Population-based cancer registries generally provide incidence rates and trends for cancer surveillance purposes, and support related clinical and epidemiological research. To receive accreditation from the COC, hospital cancer registries are required to report only in situ and primary malignant tumors for all sites, including the brain and other CNS sites.[34] Because data collection for nonmalignant intracranial and CNS tumors is not standardized at either the central tumors can be attributed to heredity or hospital registry level, collecting data for all primary intracranial tumors is a challenge.

In 1997, CBTRUS conducted a state survey (Appendix A) of benign brain and other CNS tumor collection.[35] This survey gathered information from 64 member registries of the North American Association of Central Cancer Registries (NAACCR) representing 48 states. All 64 of the registries collected data for primary malignant brain and other CNS tumors; however, only 15 state registries collected data for nonmalignant (i.e., benign) tumors at the time of the survey. Seven state registries had stopped collecting data for benign brain and other CNS tumors, three collected the data passively, one had started to collect the data in 1997, and one planned to start in 1998.The majority of the registries surveyed indicated that the requirements of their brain and other CNS tumor data collection had not been altered in the past 5 years.

METHODS

To analyze available data on issues related to the collection of data for primary intracranial and CNS tumors, the BTWG examined surveillance data from CBTRUS; the Minnesota Cancer Surveillance System (MCSS); NCDB; and SEER (Table 1). The NCDB includes cases from all 50 states, the District of Columbia, and Puerto Rico; however, it is not population-based. CBTRUS, MCSS and the SEER Program are population-based registries that cover selected areas of the U.S. Three SEER areas collect information for benign and uncertain (i.e., nonmalignant) brain tumors, but they do not report it. Currently, however, there are no standard definitions for the collection of site and type among the areas. CBTRUS, MCSS, and NCDB collect information on both malignant and nonmalignant primary brain tumors. Because the accessioning of nonmalignant cases for NCDB is not required by COC, only 59 percent of the hospitals that submitted any data to NCDB on intracranial/CNS cases diagnosed during 1989-1994 submitted nonmalignant cases. The NCDB data reported here are restricted to those hospitals that reported at least one nonmalignant case as these cases were thought to contribute most toward the purpose of this report. Also, although MCSS collects data for both malignant and nonmalignant tumors, it does so for microscopically-confirmed cases only. This registry performs 100-percent case finding audits in all pathology laboratories in Minnesota.

Data inclusion criteria were developed as specified in Appendix B, and all four of the resources (i.e., CBTRUS, MCSS, NCDB, SEER) developed subsets of their currently available data according to the agreed-upon inclusion criteria. Frequencies and proportions were computed to reflect variations in factors other than site, and the resulting contrasts across these registries broadly reflect data collection practices. For example, the primary difference in brain and CNS definition between SEER and CBTRUS lies in reporting by behavior, with the latter including all nonmalignant tumors. As such, differences in proportions between these two registries approximately reflect the magnitude of nonmalignant brain and CNS tumors. Similarly, the primary difference in brain and CNS definition between MCSS and CBTRUS is that the former requires microscopic confirmation of all tumors and the latter includes clinically and radiologically diagnosed tumors. Another important difference

between MCSS and CBTRUS is that MCSS uses a more active method for case finding. Therefore, differences in proportions reported reflect the interaction between the magnitude of nonmicroscopically diagnosed tumors and the magnitude of incomplete case finding of certain tumor types in CBTRUS. Finally, the primary difference between NCDB and CBTRUS is that the former is a hospital-based reporting system and the latter is a population-based reporting system. Therefore, differences in proportions may largely reflect referral biases inherent in hospital reporting systems. While other data collection practices may influence information in these data resources, these data bases were selected to highlight the variation in current information on primary brain and CNS tumors that arises without a standard definition.

Frequencies were computed using ICD-O codes for topography (i.e., location), behavior, diagnostic confirmation, histology, and selected combinations of behavior and topography codes. Topography was broadly defined into two main categories with subcategories as shown in Appendix B. Intracranial tumors included tumors located in the brain and other CNS regions (including the cranial nerves), in the meninges, and in the pituitary and pineal glands. Extracranial tumors included those located in the spinal cord and spinal meninges. The distinction between intracranial and extracranial tumors was removed for Table 7 by collapsing brain and spinal meninges into one category, and combining the pituitary and pineal glands into one group (as these regions have endocrine functions) to allow for adequate numbers within categories of behavior. For Table 10 the intracranial and extracranial distinction was maintained; however, the pituitary and pineal glands were collapsed into one category and the spinal cord and spinal meninges were collapsed into the extracranial category.

Behavior was coded using ICD-O categories 0 for benign, 1 for uncertain, and 3 for malignant, with the first two of these categories collapsed into a nonmalignant category. Microscopic confirmation is based on the SEER definition and grouped into positive microscopic confirmation, radiography without microscopy, clinical, and unknown. These categories were further grouped as microscopically confirmed and as not microscopically confirmed for the analysis in Table 3.

CBTRUS developed preliminary histology groupings with the aim of improved clinical relevance. ICD-O morphology codes, which are used by cancer registries, were grouped based on WHO categories for brain tumors. The details of these groupings are shown in Appendix C.

RESULTS

Percentages of Primary Intracranial and CNS Tumors by Tumor Location and by Microscopic Confirmation

Intracranial tumors comprise more than 94 percent of primary intracranial and CNS tumors, and between 50 and 92 percent of primary intracranial and CNS tumors occur in the brain (Table 2). The data in this report also show that a slightly greater percentage of malignant intracranial tumors, including malignant brain tumors, are microscopically confirmed compared with corresponding tumors of benign or uncertain behavior (Table 3). However, regardless of tumor behavior, the percentages of extracranial CNS tumors that are microscopically confirmed are higher than the percentages of intracranial tumors that are confirmed by pathologists (Table 3). Few primary intracranial and CNS tumors are not microscopically confirmed; approximately 11 percent of diagnoses are based only on a clinical or radiographical (without microscopy) assessment (Table 4).

Percentages of Malignant and of Nonmalignant Intracranial and CNS Tumors by Location

The relative distribution of nonmalignant intracranial and CNS tumors varies by primary site. While greater than 90 percent of malignant intracranial and CNS tumors occur in the brain (Table 5), only 9 to 26 percent of the nonmalignant tumors occur as parenchymal tumors (Table 6). Nonmalignant tumors comprised a significant portion of the primary intracranial and CNS tumors reported by the sources. The ratios of nonmalignant to primary malignant intracranial and CNS tumors reported by CBTRUS, MCSS, and NCDB were 0.9, 1.0, and 0.5, respectively (Table 7).

Percentages of Malignant and of Nonmalignant Intracranial and CNS Tumors by Histology

Among the malignant primary intracranial and CNS tumors, neuroepithelial (i.e., astrocytic) tumors represent 83 to 90 percent of the cases depending on the data source;

lymphomas/hemopoietic tumors, 6 to 11 percent; tumors of the meninges, 1 to 2 percent; and tumors of cranial and spinal nerves, 0.2 percent (Table 8). For nonmalignant primary intracranial and CNS tumors, tumors of the meninges represent 48 to 59 percent of the cases; tumors of the sellar region (including pituitary tumors and craniopharyngiomas), 21 to 26 percent; tumors of cranial and spinal nerves, 11 to 20 percent; and neuroepithelial tumors, 4 to 5 percent (Table 9).

Percentages of Primary Intracranial and CNS Tumors by Method of Case Finding (MCSS)

Case finding audits by the MCSS occur on an annual basis in all pathology laboratories in Minnesota and include the review of surgery/pathology, cytology, autopsy, and hematology records. These audits enable the registry to ensure that there is complete reporting of cases of malignant and nonmalignant tumors. In tables 10 through 13, data from MCSS on primary intracranial and CNS tumors are presented by method of case finding. The two methods of case finding are "routine reporting" and "special efforts." A case was found by routine reporting if one or more reports received before the close-out date for a given diagnosis year was initiated by a hospital or nonhospital facility. In contrast, a case was found by special efforts if all reports of that case received before the close-out date were requested as a result of pathology laboratory audits. The data in Table 10 indicate that routine reporting of nonmalignant tumors was less complete compared with reporting for malignant tumors. The percentages of intracranial tumor cases that were found by special efforts have varied from year to year, but the trend suggests that routine reporting may be improving (Table 11). The greatest improvement has been in the coverage of nonbrain parenchymal tumors (i.e., pituitary/pineal and other intracranial). The small numbers of autopsy-only tumors were more likely to be found by special case finding efforts than by other methods of microscopic confirmation (data not shown).

Most hospitals in Minnesota have cancer registries through which their tumor cases are reported. However, some of the cases that are seen at hospitals with registries are reported only through pathology laboratories affiliated with the hospitals. Approximately 80 percent (1451/1776) of the cases of nonmalignant intracranial tumors in the MCSS data base were reported by hospitals with registries (Table 12); eighty-five percent (1226/1451) of these cases were reported by the hospitals' registries (Table 13).The remaining 15 percent (225/1451) of the

cases were reported through the pathology laboratories affiliated with the hospitals with registries (i.e., they were not accessioned by the registries) (Table 13). The percentages of cases of nonmalignant tumors that were reported only by the pathology laboratories at facilities having cancer registries can be viewed as a measure of how frequently the registries failed to accession the cases. Of the 225 nonaccessioned tumors, half would have been missed without the MCSS case finding audits of pathology laboratories (Table 13).

Percentages of Hospital Cancer Registries That Accessioned Malignant and Nonmalignant Intracranial and CNS Tumors (NCDB)

Based on data from NCDB, more than 1500 hospitals accessioned cases with either malignant or nonmalignant intracranial and CNS tumors, and approximately 800 hospitals accessioned extracranial cases. Cases with tumors that involved the cranial or spinal meninges or the pituitary gland were more likely to be reported by hospitals that accessioned nonmalignant primary intracranial and CNS tumors than by hospitals that accessioned only malignant tumors (Table 14).

DISCUSSION

The findings for this report indicate that collection and reporting of incidence data for primary malignant intracranial and other CNS tumors are well standardized. However, substantial variation exists in the processes of collecting and reporting nonmalignant tumor data. One obstacle is that not all hospitals have cancer registries. Another problem is that not all hospitals with registries accession cases of nonmalignant tumors. Data from Minnesota (Table 13) suggest that approximately 85 percent of cases of nonmalignant intracranial tumors are accessioned by hospital registries. However, nonaccessioned CNS cases are less likely to be routinely reported to the central registry (Table 13). These differences in collection and reporting practices among the registries make it difficult to assess the burden of primary intracranial and CNS tumors.

Terminology for describing intracranial and CNS tumors is also not standardized. For example, although the pituitary gland and pineal glands are not technically part of the CNS, tumors that involve these organs are often included in the term "CNS tumors." On the other hand, some consider the eye to be intracranial; however, "CNS tumors" generally do not include the

eyes. Although the World Health Organization (WHO) has published a list of histologic groupings, at present no single comprehensive list of histologic groupings exists for intracranial and CNS tumors. Since the International Classification of Childhood Cancer (ICCC) is based on histologic type rather than site, certain benign histologies of sites other than brain/CNS (e.g., non-CNS ependymomas) would not be included in the BTWG's primary intracranial and CNS histology groupings (Appendix D). Thus, the variation in incidence estimates of primary intracranial and CNS tumors may be attributable, in part, to the variation in the definitions of these tumors. In addition, differences in cancer registry training and procedures may contribute to nonreporting and inconsistent reporting of the tumors.

The BTWG discussed the feasibility of conducting a random sampling procedure as an alternative to complete enumeration and ongoing surveillance of nonmalignant brain tumors. It was agreed that this technique would not be valid or feasible for the following reasons:

1. If a random sample of incident brain tumors were desired, a complete list would be needed first (a sampling frame is required).
2. If a survey method were used to identify brain tumors in the general population (e.g., random digit dialing or other general population survey techniques), the information needed could not be obtained for several reasons:
    - Either a prohibitively large numbers of individuals would have to be surveyed, or else only a very small number of brain tumors could be identified, given the rarity and sometimes short survival time.
    - The brain tumors identified would not be representative of all brain tumors that occur, because people who had survived their brain tumors would be more likely to be identified.
    - The incidence rate would still not be known, since cross-sectional surveys only identify disease prevalence.
3. If a sample of hospitals were selected to contribute data, the tumors included would not represent the entire spectrum of brain tumors because those that were diagnosed and treated outside of the hospital setting would be excluded.

An alternative approach to sampling, and the least troublesome, would be to conduct population-based surveillance only in certain geographic areas of the country. If one assumes that the etiology of brain tumors does not differ from one area of the country to another, then the scientific validity of studies based on this type of sampling would not be a problem. However, given the rarity of some types of brain tumors, sufficient numbers for study may not be available unless the vast majority of the nation's population were covered by population-based data collection. Also, geographic areas without population-based surveillance would have no data to use for assessing local trends or variations in brain tumor occurrence.

In the absence of standard tumor registration procedures and training, special case finding efforts by central registries may be necessary to ensure that complete data are collected for nonmalignant intracranial and CNS tumors. Data from MCSS (Table 10), which has a legal requirement for reporting nonmalignant intracranial tumors and performs 100 percent case finding audits in all laboratories, indicate that only 80 percent of nonmalignant tumors were routinely reported. In contrast, routine reporting accounted for more than 95 percent of malignant intracranial and CNS tumors. Nonmalignant cases in Minnesota that were not accessioned by hospital registries had a 50 percent chance of being routinely reported to the central registry; however, these were only microscopically confirmed cases.

Completeness of reporting is critical to cancer registries. Accurate case counts are necessary to assess the burden of cancer, to guide cancer control program planning, to prioritize the allocation of health resources, and to facilitate epidemiologic research. Most central cancer registries have state laws that mandate reporting of cases by physicians, and by hospitals, laboratories, and other facilities that provide screening, diagnostic, or therapeutic services. Complete reporting of nonmalignant intracranial and CNS tumors would be greatly improved if reporting requirements of the COC, central cancer registries, SEER, and the National Program of Cancer Registries (NPCR) were changed to require the collection of information for these tumors. However, this requirement would have many implications, including an increase in work load as well as associated costs for reporting facilities. An estimated 1.4 percent of all new cancer cases diagnosed in 1998 will involve invasive brain and other nervous system tumors;[7] since the numbers of benign and invasive brain and other CNS tumors

diagnosed annually are similar, facilities that presently do not accession nonmalignant cases could expect an approximately 1.4 percent increase in the total number of CNS cases collected by the registry. Some estimates of the percentage of brain tumors have been lower (0.5 percent)[36] and others have been higher (9 percent);[37] therefore, the extent to which the workload would be affected may vary. The data in this report suggest that if reporting of nonmalignant intracranial and CNS tumors were required, the total number of cases of tumors for these anatomic sites would double for facilities that presently report only malignant tumors. This is consistent with the findings of Davis et al,[36] who reported that the incorporation of benign brain tumors into the cancer-reporting systems of four central registries increased the overall incidence of brain cancer by 49 percent. Forty percent (913) of the hospital registries that submitted data to the NCDB did not report nonmalignant brain tumors. These registries could expect up to a 50 percent increase in the number of intracranial and CNS tumors reported; nonregistry facilities, which represent approximately 20 percent of cases reported to central cancer registries, could expect a similar increase. Also, central cancer registries that currently do not require reporting of nonmalignant intracranial and CNS tumors could expect increases in their workloads as a result of the additional time spent on processing and quality control procedures and training. The lack of standard definitions and collection and reporting guidelines would make these tasks more time-consuming as well.

For some central cancer registries that want to expand their reporting requirements to include nonmalignant intracranial and CNS tumors, a change in current legislation and/or regulations may be needed. These changes could involve several months of lead time if public hearings or other legal procedures are necessary. In other states, the reporting of these tumors is not required in either their legislation or regulations; in these situations hospitals have been asked to voluntarily report the cases. For example, in Massachusetts the reporting of benign brain tumors is not required by law, but hospitals have been reporting these cases since 1982. However, the completeness of data reported on a voluntary basis is difficult to assess. For public health surveillance systems, a mandate to report is the basic requirement of a comprehensive, higher quality system.

If COC, SEER, NPCR, and central cancer registries changed their reporting requirements to include nonmalignant intracranial and CNS tumors, the registry manuals disseminated by these programs and registries would also need to be modified to include new definitions of reportable diagnoses. All reporting facilities would have to be notified of the new requirements. To ensure complete reporting from registries, additional training of registrars should be expected. Case finding methods may need to be modified in order to identify the new cases and sources of information. Central cancer registries would also need to increase their case finding audits. The inclusion of nonmalignant tumors would also necessitate changes to other data items such as sequence number. Hospital and central cancer registries would need to consider adding WHO brain tumor grade as an additional data item.

Finally, the software used by reporting facilities and by central registries would need to be modified to accept nonmalignant morphology and behavior codes. Also, edits programs would need to be modified to include these additions and to accept a nonmalignant sequence numbering procedure.

Before reporting requirements for primary intracranial and CNS tumors could be changed, feasibility studies would have to be conducted to determine whether such changes should be recommended. Two SEER special studies are under way to evaluate the impact of requiring the collection of benign brain tumors on case finding, cost, quality control, and training. However, additional studies may be required.

RECOMMENDATIONS OF THE BRAIN TUMOR WORKING GROUP

1. **We recommend** the following standard definition for collecting precise data for all primary intracranial and central nervous system tumors:

   Primary intracranial and central nervous (CNS) tumors are primary tumors occurring in the following sites, irrespective of histologic type or behavior: brain, meninges, spinal cord, cauda equina, cranial nerves and other parts of the CNS, pituitary gland, pineal gland, and craniopharyngeal duct (see Appendix B).

2. **We recommend** the development of a standard site and histology definition for tabulating estimates of these tumors to allow comparability of information across registries. Pathologists, the North American Association of Central Cancer Registries (NAACCR), the Commission on Cancer (COC), the Surveillance, Epidemiology, and End Results (SEER) Program, the National Program of Cancer Registries (NPCR), and the International Agency for Research on Cancer (IARC) need to be involved in developing this standard.

3. **We recommend** collection of data for primary intracranial and extracranial CNS tumors by all registries, hospital- and central-based. This effort will necessitate a change in the COC requirements and will increase costs to the hospital-based programs. Federal funding should be allocated to supplement the additional transition and ongoing data collections costs that will be incurred by central registries. Before additional data collection is implemented, we recommend a pilot study should be conducted to assess the procedures and quality control functions needed, as well as the costs of collecting data for these tumors.

4. **We recommend** that the appropriate government and professional organizations be involved in carrying out the development and implementation of special training programs and curricula for central registry, hospital registry, and laboratory personnel as well as the development of computerized edit-checking procedures. Training for reporting and tabulating primary intracranial and CNS tumors should be offered on a regular basis.

REFERENCES

1. Davis F, Preston-Martin S. Epidemiology of brain tumors. *In* Bigner DD, McLendon RE, Bruner JM, eds. Russell and Rubinstein's pathology of tumors of the nervous system, ed 6. London, Arnold Publishing, 1996:5-45.
2. World Health Organization (WHO) (1976) International classification of diseases for oncology, ed 1. Geneva: World Health Organization.
3. World Health Organization (WHO) (1990) International classification of diseases for oncology, ed 2. Geneva: World Health Organization.
4. Kleihues P, Burger PC, Scheithauer BW. The new WHO classification of brain tumors. Brain Pathol 1993;3:255-268.
5. Thapar K, Laws ER. Tumors of the central nervous system. *In* Murphy GP, Lawrence W Jr, Lenhard RE Jr, eds. Textbook of clinical oncology, ed 2. Atlanta, American Cancer Society 1995;378-410.
6. Walker AE, Robins M, Weinfield FD. Epidemiology of brain tumors: the national survey of intracranial neoplasms. Neurol 1985;35:219-226.
7. Ries LAG, Kosary CL, Hankey BF, Miller BA, Edwards BK (eds). SEER Cancer Statistics Review, 1973-1995, National Cancer Institute. Bethesda, MD, 1998.
8. Cancer Facts & Figures - 1998. American Cancer Society, Atlanta, Georgia, 1998.
9. Davis FG, Bruner JM, Surawicz TS. The rationale for standardized registration and reporting of brain and central nervous system tumors in population-based cancer registries. Neuroepidemiol 1997;16:308-316.
10. CBTRUS (1996) First annual report 1995. Central Brain Tumor Registry of the United States. Chicago, Illinois.
11. Preston-Martin S. Epidemiology of primary CNS neoplasms. Neurol Clinics 1996;14: 273-290.
12. Greig NH, Ries LG, Yancik R, Rapoport SI. Increasing annual incidence of primary malignant brain tumors in the elderly. J Natl Cancer Inst 1991; 83:1621-1624.
13. Mao Y, Desmeules M. Semenciw RM, Hill G, Gaudette L, Wigle DT. Increasing brain cancer rates in Canada. Can Med Assoc J 1991;145:1583-1591.
14. Frelick R. Huang P, Topham A. Brain and central nervous system tumors: Delaware 1980 to 1989. Del Med J 1992;64:571-573.
15. Davis DL, Ahlbom A, Hoel D, Percy C. Is brain cancer mortality increasing in industrial countries? Am J Ind Med 1991;19:421-431.
16. Helseth A. Increasing incidence of primary central nervous system tumors in the elderly: Real increase or improved detection? J Natl Cancer Inst 1993;85:1871-1872.
17. Modan B, Wagener D, Feldman JJ, Rosenberg HM, Feinleib M. Increased mortality from brain tumors: A combined outcome of diagnostic technology and change of attitude toward the elderly. Am J Epidemiol 1992;135:1349-1357.
18. Polednak AP. Time trends in the incidence of brain and central nervous system cancers in Connecticut, J Natl Cancer Inst 1991;83:1679-1681.
19. Boyle P, Maissonneuve P, Saracci R, Muir CS. Is the increase in incidence of primary malignant brain tumors real? J Natl Cancer Inst 1990;20:1594-1596.

20. Preston-Martin S. Descriptive epidemiology of primary tumors of the brain, cranial nerves and spinal meninges in Los Angeles County. Neuroepidemiol 1989;8:283-295.
21. Helseth A, Mork SJ. Neoplasms of the central nervous system in Norway. Epidemiological characteristics of intracranial gliomas according to histology. APMIS *97* 1989;547-555.
22. Gurney JG, Davis S, Severson RK, Fang J-Y, Ross JA, Robison LL. Trends in cancer incidence among children in the United States. Cancer 1996;78:532-541.
23. Gurney JG, Ross JA, Wall DA, Bleyer WA, Severson RK, Robison LL. Infant cancer in the U.S.: histology-specific incidence trends, 1973-1992. J Pediatr Hem Onc 1997;19:428-432.
24. Nishio S, Takeshita I, Morioka T, Fukui M. Primary intracranial squamous cell carcinomas: report of two cases. Neurosurgery 1995;37:329-332.
25. Freilich RJ, Thompson SJ, Walker RW, Rosenblum MK. Adenocarcinomatous transformation of intracranial germ cell tumors. Am J Surg Pathol 1995;19:537-544.
26. Yuan S, Miller DW, Barnett GH, Hahn JF, Williams BR. Identification and characterization of human beta 2-chimaerin: association with malignant transformation in astrocytoma. Cancer Research 1995; 55:3456-3461.
27. Preston-Martin S, Mack WJ. Neoplasms of the nervous system. *In* Schoenfeld D, Fraumeni JF Jr., eds. Cancer epidemiology and prevention, ed 2. New York, Oxford University Press, 1996:1231-1281.
28. Riggs JE. Rising primary malignant brain tumor mortality in the elderly. A manifestation of differential survival. Arch of Neurol 1995;52:571-575.
29. Davis FG, Freels S, Grutsch J, Barlas S, Brem S. Survival rates in patients with primary malignant brain tumors stratified by patient age and tumor histological type: an analysis based on Surveillance, Epidemiology, and End Results (SEER) data, 1973-1991. J Neurosurg 1998;88:1-10.
30. Surawicz TS, Davis F, Freels S, Laws ER, Menck HR. Brain tumor survival: results from the National Cancer Data Base. (Accepted for publication in J Neuro-Oncol)
31. Bondy ML, Lustbader ED, Buffler PA, Schull WJ, Hardy RJ, Strong LC. Genetic epidemiology of childhood brain tumors. Genet Epidemiol 1991;8:253-267.
32. Preston-Martin S, Monroe K, Lee P, Bernstein L, Kelsey J, Henderson S, Forrester D, Henderson B. Spinal meningiomas in women in Los Angeles County: Investigation of an etiologic hypothesis. Cancer Epidemiol Biomarkers Prev 1995;4:333-339.
33. Davis JR, Brownson RC, Garcia R, Bentz BJ, Turner A. Family pesticide use and childhood brain cancer. Arch Environ Contam Toxicol 1993:24:87-92.
34. Commission on Cancer. General principles and coding. *In* Standards of the Commission on Cancer, vol. II: Registry Operations and Data Standards. American College of Surgeons. Chicago, Illinois. 1996.
35. Central Brain Tumor Registry of the United States (CBTRUS). 1997 State Survey of Benign Brain Tumor Collection. Chicago, Illinois. 1997.
36. Davis FG, Malinski N, Haenszel W, Chang J, Flannery J, Gershman S, Dibble R, Bigner DD. Primary brain tumor incidence rates in the four United States regions, 1985-1989: A pilot study. Neuroepidemiol 1996;15:103-112.
37. Schoenberg BS, Christine BW, Whisnant JP. The descriptive epidemiology of primary intracranial neoplasms: the Connecticut experience. Am J Epidemiol 1976;104:499-510.

TABLE 1. COMPARISON OF REGISTRIES SUPPLYING DATA ON PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS IN THE UNITED STATES

	Central Brain Tumor Registry of the United States (CBTRUS)	Minnesota Cancer Surveillance System (MCSS)	ACoS National Cancer Data Base (NCDB)	Surveillance, Epidemiology, and End Results Program (SEER)
Purpose	"CBTRUS centralizes population-based incidence data on all brain tumors and uses other existing data resources to characterize the incidence, mortality, and survival of patients with brain tumors."	The MCSS collects information on all cancers diagnosed since 1988 among Minnesota residents. The MCSS is population-based, pathology-based, and is an active system.	"The goal of the NCDB is to present an annual summary of patient care for cases of cancer diagnosed and treated at hospitals throughout the country."	"A continuing project of the NCI, the SEER program collects cancer data on a routine basis from designated population-based cancer registries in various areas of the country."
Target population	The populations covered by registries in **Arizona, Colorado, Connecticut, Delaware, Idaho, Maine, Massachusetts, Minnesota, Missouri, Montana, North Carolina**, and **Utah**. The CBTRUS population represents a nonrandom 15 percent sample of the U.S. population.	The population of **Minnesota.**	The populations treated at participating hospitals in **the 50 states, the District of Columbia,** and **Puerto Rico.** NCDB is not a population-based registry. At present, it is estimated to cover approximately 57 percent of total cases nationwide.	The populations covered by the state registries in **Connecticut, Hawaii, Iowa, New Mexico**, and **Utah**, as well as the regional registries in **Atlanta, Georgia; Detroit, Michigan; San Francisco-Oakland, California**; and **Seattle-Puget Sound, Washington.** The SEER population represents a nonrandom 14 percent sample of the U.S. population.

	Central Brain Tumor Registry of the United States (CBTRUS)	Minnesota Cancer Surveillance System (MCSS)	ACoS National Cancer Data Base (NCDB)	Surveillance, Epidemiology, and End Results Program (SEER)
Type of primary intracranial and central nervous system tumors reported	Data on newly-diagnosed cases of benign and malignant primary brain tumors based on ICD-O codes.	Data on newly-diagnosed, microscopically-confirmed tumors of the central nervous system, including pituitary and pineal glands.	Hospitals voluntarily submit full analytic cancer registry caseloads with data items and codes defined by the Commission on Cancer (COC) for hospital cancer registries. Most participating hospitals have programs approved by COC or are establishing program approval. NCDB calls for data do not limit cases by malignant behavior, but accessioning of non-malignant cases is not required for program approval. Fifty-nine percent of the hospitals that submitted any intracranial/CNS cases diagnosed during 1989-1994 submitted at least one nonmalignant case. The NCDB data in this report are limited to the cases submitted by these hospitals (82 percent of all intracranial and extracranial central nervous system cases on file).	Trends in the incidence, mortality, and patient survival of brain tumors in the U.S. However, SEER does not collect information on benign and uncertain brain tumors.
Other comments	Duplicate cancer registrations can not be identified by the participating registries.	The MCSS performs 100 percent case finding audits in all pathology laboratories on an annual basis. Therefore, the data from the MCSS might be considered a “best-case scenario” in terms of completeness of reporting for microscopically-confirmed tumors.	Report duplication for 1989-1994 intracranial and extracranial central nervous system reports was 6.7 percent. However, the NCDB data in Tables 2 through 9 are based on unduplicated reports.	

TABLE 2. NUMBER AND PERCENT OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS BY TUMOR LOCATION

Location[a]	CBTRUS[b] Number (%)	MCSS[c] Number (%)	NCDB[d] Number (%)	SEER[e] Number (%)
Intracranial	13,084 (94.7)	3,600 (93.6)	66,322 (95.5)	10,196 (96.9)
Brain	7,999 (57.9)	1,924 (50.0)	48,527 (69.9)	9,721 (92.4)
Meninges	2,719 (19.7)	838 (21.8)	9,365 (13.5)	139 (1.3)
Pituitary/ craniopharygeal duct	1,491 (10.8)	530 (13.8)	4,958 (7.1)	30 (0.3)
Pineal	74 (0.5)	21 (0.5)	477 (0.7)	80 (0.8)
Other central nervous system, including cranial nerves	801 (5.8)	287 (7.5)	2,995 (4.3)	226 (2.2)
Extracranial	730 (5.3)	245 (6.4)	3,137 (4.5)	322 (3.1)
Spinal cord	497 (3.6)	47 (3.8)	2,395 (3.4)	313 (3.0)
Spinal meninges	233 (1.7)	98 (2.5)	742 (1.1)	9 (0.1)
TOTAL	13,814 (100)	3,845 (100)	69,459 (100)	10,518 (100)

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993.

[a]Refer to the sites in Appendix B
[b]Central Brain Tumor Registry of the United States
[c]Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[e]Survival, Epidemiology, and End results Program (malignant cases only).

TABLE 3. ROW PERCENT DISTRIBUTION OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS BY TUMOR BEHAVIOR AND MICROSCOPIC CONFIRMATION

Location[a]	CBTRUS[b]			MCSS[c]			NCDB[d]			SEER[e]		
	Total	Percent Micro. Confirmed	Percent Not Micro. Confirmed	Total	Percent Micro. Confirmed	Percent Not Micro. Confirmed	Total	Percent Micro. Confirmed	Percent Not Micro. Confirmed	Total	Percent Micro. Confirmed	Percent Not Micro. Confirmed
Intracranial	13,084	86.7	13.3	3,600	100	NA	66,322	88.7	11.3	10,196	87.1	12.9
Malignant	7,171	89.2	10.8	1,824	100	NA	44,801	91.3	8.7	10,196	87.1	12.9
Benign and uncertain	5,913	83.5	16.5	1,776	100	NA	21,521	83.2	16.8	NA	NA	NA
Brain	7,999	88.2	11.8	1,924	100	NA	48,527	90.2	9.8	9,721	87.3	12.7
Malignant	6,862	89.6	10.4	1,746	100	NA	42,622	91.5	8.5	9,721	87.3	12.7
Benign and uncertain	1,137	80.6	19.4	178	100	NA	5,905	80.6	19.4	NA	NA	NA
Extracranial	730	96.2	3.8	245	100	NA	3,137	96.0	4.0	322	95.7	4.3
Malignant	242	93.4	6.6	60	100	NA	1,483	96.6	3.4	322	95.7	4.3
Benign and uncertain	488	97.5	2.5	185	100	NA	1,654	95.5	4.5	NA	NA	NA
TOTAL[f]	13,814	87.2	12.8	3,845	100	NA	69,459	89.0	11.0	10,518	87.4	12.6
Malignant	7,413	89.4	10.6	1,884	100	NA	46,284	91.5	8.5	10,518	87.4	12.6
Benign and uncertain	6,401	84.6	15.4	1,961	100	NA	23,175	84.1	15.9	NA	NA	NA

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993.

[a]Refer to the site codes in Appendix B.
[b]Central Brain Tumor Registry of the United States.
[c]Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[e]Surveillance, Epidemiology, and End Results Program (malignant cases only).
[f]Includes intracranial and extracranial cases.
NA = Not applicable.

TABLE 4. NUMBER AND PERCENT OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS BY DIAGNOSTIC CONFIRMATION

Diagnostic Confirmation	CBTRUS[a] Number (%)	MCSS[b] Number (%)	NCDB[c] Number (%)	SEER[d] Number (%)
Positive microscopy	12,087 (87.5)	3,845 (100)	61,818 (89.0)	9,188 (87.4)
Radiography without microscopy	1,306 (9.5)	NA	6,759 (9.7)	1,015 (9.7)
Clinical	204 (1.5)	NA	484 (0.7)	98 (0.9)
Unknown	217 (1.5)	NA	398 (0.6)	217 (2.1)
TOTAL	13,814 (100)	3,845 (100)	69,459 (100)	10,518 (100)

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993.

[a]Central Brain Tumor Registry of the United States.
[b] Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[c]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[d]Surveillance, Epidemiology, and End Results Program (malignant cases only).
NA = Not applicable

TABLE 5. NUMBER AND PERCENT OF MALIGNANT PRIMARY INTRACRANIAL AND CENTRAL SYSTEM TUMORS BY TUMOR LOCATION

Location[a]	CBTRUS[b] Number (%)	MCSS[c] Number (%)	NCDB[d] Number (%)	SEER[e] Number (%)
Intracranial	7,171 (97.7)	1,824 (96.8)	44,801 (96.8)	10,196 (96.9)
Brain	6,862 (92.6)	1,746 (92.7)	42,622 (92.1)	9,721 (92.4)
Meninges	98 (1.3)	23 (1.2)	581 (1.3)	139 (1.3)
Pituitary/ craniopharyngeal duct	37 (0.5)	13 (0.7)	189 (0.4)	30 (0.3)
Pineal	58 (0.8)	20 (1.1)	378 (0.8)	80 (0.8)
Other central nervous system, including cranial nerves	116 (1.6)	22 (1.2)	1,031 (2.2)	226 (2.1)
Extracranial	242 (3.3)	60 (3.2)	1,483 (3.2)	322 (3.1)
Spinal cord	234 (3.2)	60 (3.2)	1,410 (3.0)	313 (3.0)
Spinal meninges	8 (0.1)	0	73 (0.2)	9 (0.1)
TOTAL	7,413 (100)	1,884 (100)	46,284 (100)	10,518 (100)

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993.

[a]Refer to the site codes in Appendix B.
[b]Central Brain Tumor Registry of the United States
[c]Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[e]Survival, Epidemiology, and End Results Program (malignant cases only).

TABLE 6. NUMBER AND PERCENT OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS OF BENIGN AND UNCERTAIN BEHAVIOR BY TUMOR LOCATION

Location[a]	CBTRUS[b] Number (%)	MCSS[c] Number (%)	NCDB[d] Number (%)	SEER[e] Number (%)
Intracranial	5,913 (92.4)	1,776 (90.6)	21,521 (92.9)	NA
Brain	1,137 (17.8)	178 (9.1)	5,905 (25.5)	NA
Meninges	2,621 (40.9)	815 (41.6)	8,784 (37.9)	NA
Pituitary/ craniopharyngeal duct	1,454 (22.7)	517 (26.4)	4,769 (20.6)	NA
Pineal	16 (0.2)	1 (<0.1)	99 (0.4)	NA
Other central nervous system, including cranial nerves	685 (10.7)	265 (13.5)	1,964 (8.5)	NA
Extracranial	488 (7.6)	185 (9.4)	1,654 (7.1)	NA
Spinal cord	263 (4.1)	87 (4.4)	985 (4.2)	NA
Spinal meninges	225 (3.5)	98 (5.0)	669 (2.9)	NA
TOTAL	6,401 (100)	1,961 (100)	23,175 (100)	NA

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993.

[a]Refer to the site codes in Appendix B.
[b]Central Brain Tumor Registry of the United States
[c]Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[e]Surveillance, Epidemiology, and End Results Program (malignant cases only).
NA = Not applicable.

TABLE 7. NUMBER AND PERCENT OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS BY TUMOR BEHAVIOR AND LOCATION

Behavior/ Tumor Location[a]	CBTRUS[b] Number (%)	MCSS[c] Number (%)	NCDB[d] Number (%)	SEER[e] Number (%)
Malignant	7,413 (53.7)	1,884 (49.0)	46,284 (66.6)	10,518 (100)
Brain	6,862 (49.7)	1,746 (45.4)	42,622 (61.4)	9,721 (92.4)
Meninges	98 (0.7)	23 (0.6)	581 (0.8)	139 (1.3)
Pituitary/craniopharyngeal duct/pineal	95 (0.7)	33 (0.9)	567 (0.8)	110 (1.1)
Other central nervous system including cranial nerves	116 (0.8)	22 (0.6)	1,031 (1.5)	226 (2.2)
Spinal cord	242 (1.8)	60 (1.6)	1,483 (2.1)	322 (3.1)
Benign	5,794 (41.9)	1,787 (46.5)	20,694 (29.8)	NA
Brain	803 (5.8)	99 (2.6)	4,491 (6.5)	NA
Meninges	2,565 (18.6)	807 (21.0)	8,613 (12.4)	NA
Pituitary/craniopharyngeal duct/pineal	1,322 (9.6)	458 (11.9)	4,330 (6.2)	NA
Other central nervous system including cranial nerves	678 (4.9)	263 (6.8)	1,911 (2.8)	NA
Spinal cord	426 (3.1)	160 (4.2)	1,349 (1.9)	NA
Uncertain	607 (4.4)	174 (4.5)	2,481 (3.6)	NA
Brain	334 (2.4)	79 (2.1)	1,414 (2.0)	NA
Meninges	56 (0.4)	8 (0.2)	171 (0.2)	NA
Pituitary/craniopharyngeal duct/pineal	148 (1.1)	60 (1.6)	538 (0.8)	NA
Other central nervous system including cranial nerves	7 (0.1)	2 (0.1)	53 (0.1)	NA
Spinal cord	62 (0.5)	25 (0.7)	305 (0.4)	NA
TOTAL	13,814 (100)	3,845 (100)	69,459 (100)	10,518 (100)

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993. NA=Not applicable.
[a]Refer to the site codes in Appendix B; [b]Central Brain Tumor Registry of the United States;
[c]Minnesota Cancer Surveillance System (microscopically confirmed cases only);
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases; [e]Survival, Epidemiology, and End Results Program (malignant cases only).

TABLE 8. NUMBER AND PERCENT OF MALIGNANT PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS BY HISTOLOGY GROUPINGS

WHO Histology[a] Groupings	CBTRUS[b] Number (%)	MCSS[c] Number (%)	NCDB[d] Number (%)	SEER[e] Number (%)
Neuroepithelial	6,477 (87.4)	1,689 (89.6)	39,701 (85.8)	8,676 (82.5)
Tumors of cranial and spinal nerves	12 (0.2)	3 (0.2)	108 (0.2)	25 (0.2)
Tumors of meninges	137 (1.8)	25 (1.3)	994 (2.1)	194 (1.8)
Lymphomas/ Hemopoietic	473 (6.4)	121 (6.4)	3,492 (7.5)	1,099 (10.5)
Germ cell	70 (0.9)	20 (1.1)	403 (0.9)	97 (0.9)
Cysts and tumor-like lesions	0	0	0	0
Tumors of sellar region	20 (0.3)	11 (0.6)	100 (0.2)	6 (0.1)
Local extensions	29 (0.4)	11 (0.6)	166 (0.4)	38 (0.4)
Unclassified/ unassigned tumors[f]	195 (2.6)	4 (0.2)	1,320 (2.9)	383 (3.6)
TOTAL	7,413 (100)	1,884 (100)	46,284 (100)	10,518 (100)

Note: Reported data are incident cases from 1989-1994 except for CBTRUS data, which are from 1990-1993.

[a]Refer to the World Health Organization (WHO) histology groupings in Appendix C.
[b]Central Brain Tumor Registry of the United States
[c]Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[f]For MCSS, unclassified/unassigned tumors include one malignant and four nonmalignant histologies which were not listed in the CBTRUS classification scheme. For NCDB, unclassified/unassigned tumors include 319 cases (less than one half percent) which had histologies not assigned by CBTRUS to any other category. For SEER, unclassified/unassigned tumors include all malignant histologies occurring in the brain/CNS which could not be classified in one of the above categories based on their 4-digit histologic type.

TABLE 9. NUMBER AND PERCENT OF PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS OF BENIGN AND UNCERTAIN BEHAVIOR BY HISTOLOGY GROUPINGS

WHO Histology[a] Groupings	CBTRUS[b] Number (%)	MCSS[c] Number (%)	NCDB[d] Number (%)	SEER[e] Number (%)
Neuroepithelial	275 (4.3)	91 (4.6)	1,064 (4.6)	NA
Tumors of cranial and spinal nerves	980 (15.3)	397 (20.2)	2,618 (11.3)	NA
Tumors of meninges	3,509 (54.8)	941 (48.0)	13,667 (59.0)	NA
Lymphomas/ Hemopoietic	3 (<0.1)	0	7 (<0.1)	NA
Germ cell	4 (<0.1)	1 (0.1)	25 (0.1)	NA
Cysts and tumor-like lesions	15 (0.2)	3 (0.2)	34 (0.1)	NA
Tumors of sellar region	1,444 (22.6)	510 (26.0)	4,749 (20.5)	NA
Local extensions	0	0	0	NA
Unclassified/ unassigned tumors[f]	171 (2.7)	18 (0.9)	1,011 (4.4)	NA
TOTAL	6,401 (100)	1,961 (100)	23,175 (100)	NA

Note: Reported data are incident cases from 1989-1994 except for CBTRUS, which is 1990-1993.

[a]Refer to the World Health Organization (WHO) histology groupings in Appendix C.
[b]Central Brain Tumor Registry of the United States
[c] Minnesota Cancer Surveillance System (microscopically confirmed cases only).
[d]National Cancer Data Base (limited to cases reported by hospitals that accessioned nonmalignant cases).
[e]Survival, Epidemiology, and End Results Program (malignant cases only).
[d]For MCSS, unclassified/unassigned tumors include one malignant and four nonmalignant histologies which were not listed in the CBTRUS classification scheme. For NCDB, unclassified/unassigned tumors include 319 cases (less than one half percent) which had histologies not assigned by CBTRUS to any other category. For SEER, unclassified/unassigned tumors include all malignant histologies occurring in the brain/CNS which could not be classified in one of the above categories based on their 4-digit histologic type.
NA = Not applicable.

TABLE 10. METHOD OF CASE FINDING FOR PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS BY TUMOR BEHAVIOR AND LOCATION, MINNESOTA, 1989-1994

Behavior/ Tumor Location[a]	Number of Cases	Percent Found by Routine Reporting	Percent Found by Special Efforts
Malignant	1,884	96.1	3.9
Brain	1,746	96.5	3.5
Cerebral meninges	23	95.7	4.3
Pituitary/cranio. duct/ pineal	33	93.9	6.1
Other intracranial	22	86.4	13.6
Extracranial	60	88.3	11.7
Benign	1,787	78.8	21.2
Brain	99	83.8	16.2
Cerebral meninges	807	84.3	15.7
Pituitary/cranio. duct/ pineal	458	74.0	26.0
Other intracranial	263	77.9	22.1
Extracranial	160	63.1	36.9
Uncertain	174	85.6	14.4
Brain	79	82.3	17.7
Cerebral meninges	8	87.5	12.5
Pituitary/craniopharyn-geal duct/pineal	60	85.0	15.0
Other intracranial	2	100.0	0.0
Extracranial	25	96.0	4.0
TOTAL	3,845	87.6	12.4

[a]Refer to the site codes in Appendix B.

TABLE 11. METHOD OF CASE FINDING FOR INTRACRANIAL[a] TUMORS BY DIAGNOSIS YEAR, MINNESOTA, 1989-1994

Diagnosis Year	Number of Cases	Percent Found by Routine Reporting	Percent Found by Special Efforts
1989	571	83.7	16.3
1990	616	86.5	13.5
1991	586	91.3	8.7
1992	602	89.9	10.1
1993	633	87.2	12.8
1994	592	92.9	7.1
TOTAL	3,600	88.6	11.4

[a]Includes the brain, cerebral meninges, pituitary gland, pineal gland, and other intracranial tumors (refer to the site codes in Appendix B).

TABLE 12. METHOD OF CASE FINDING FOR NONMALIGNANT INTRACRANIAL[a] TUMORS BY TYPE OF FACILITY, MINNESOTA, 1989-1994

Type of Facility	Number of Tumors	Percent Found by Routine Reporting	Percent Found by Special Efforts
Registry at facility	1,451	85.9	14.1
No registry at facility	325	56.9	43.1
TOTAL	1,776	80.6	19.4

[a]Includes tumors with benign or uncertain behavior that involve the brain, cerebral meninges, pituitary gland, pineal gland, or other intracranial sites (refer to the site codes in Appendix B).

TABLE 13. REPORTING METHOD AND SOURCE OF CASE FINDING FOR NON-MALIGNANT INTRACRANIAL[a] TUMORS DIAGNOSED IN HOSPITALS WITH A TUMOR REGISTRY, MINNESOTA, 1989-1994

Reporting Method	Number of Tumors	Percent Found by Routine Reporting	Percent Found by Special Efforts
Registry-reported	1,226	92.5	7.5
Laboratory-reported	225	50.2	49.8
TOTAL	1,451	85.9	14.1

[a]Includes tumors with benign or uncertain behavior that involve the brain, cerebral meninges, pituitary gland, pineal gland, or other intracranial sites (refer to the site codes in Appendix B).

TABLE 14. HOSPITAL REGISTRY ACCESSIONING OF MALIGNANT AND NON-MALIGNANT PRIMARY INTRACRANIAL AND CENTRAL NERVOUS SYSTEM TUMORS, NCDB, 1989-1994

	BEHAVIOR					
	TUMOR		Malignant [c]		Nonmalignant[d]	
	Total[b]					
Tumor Location[a]	Hospitals Reporting		Hospitals Reporting		Hospitals Reporting	
	Number	(%)	Number	(%)	Number	(%)
Intracranial	1,542	(100)	1,526	(100)	900	(99)
Brain	1,527	(99)	1,519	(>99)	754	(83)
Meninges	740	(48)	390	(26)	613	(67)
Pituitary gland/craniopharyn-geal duct	602	(39)	168	(11)	538	(59)
Pineal gland	319	(21)	277	(18)	79	(9)
Other central nervous system, including cranial nerves	648	(42)	545	(36)	299	(33)
Extracranial	812	(53)	708	(46)	410	(45)
Spinal cord	760	(49)	692	(45)	311	(34)
Spinal meninges	299	(19)	72	(5)	258	(28)
TOTAL Hospitals	1,542	(100)	1,526	(100)	913	(100)

Note: The number of hospitals in each of the second, third, and fourth columns is the number that submitted reports to the NCDB for any intracranial and central nervous system tumors for the respective malignancy status. Not all hospitals submitted reports for each year during the study period (1989-1994).

[a]Refer to the site codes in Appendix B.
[b]Behavior codes = 0, 1, 3
[c]Behavior code = 3.
[d]Behavior codes = 0, 1

APPENDIX A

1997 CBTRUS State Survey of Benign Brain Tumor Data Collection

State	Collection Status
Currently Collecting Benign Tumor Data	
1. Arizona Cancer Registry	since 1981
2. Colorado Central Cancer Registry	since 1988
3. Connecticut Tumor Registry	since 1965
4. Idaho Cancer Data Registry	since 1970
5. Maine Cancer Registry	since 1983
6. Massachusetts Cancer Registry	since 1982
7. Minnesota Cancer Surveillance System	since 1988
8. Montana Central Tumor Registry	since 1979
9. New York State Cancer Registry	since 1988
10. North Carolina Central Cancer Registry	since 1990
11. Utah Cancer Registry	since 1966
12. Virginia Cancer Registry	since 1994
13. Delaware State Cancer Registry	
14. Washington State Cancer Registry	since 1992
15. Texas Cancer Registry	passive collection 1979-1994 active collection since 1995
Stopped or Passive Collection of Benign Tumor Data	
1. New Jersey State Cancer Registry	stopped 1979-1995
2. Kansas Cancer Registry	stopped 1969-1995
3. New Hampshire State Cancer Registry	stopped 1986-1991
4. District of Columbia Cancer Registry	stopped 1997
5. Wyoming Cancer Surveillance Program	stopped 1/1/1996
6. Louisiana Tumor Registry	stopped 1992 (passive collection)
7. Illinois State Cancer Registry	stopped 1/1/1996
8. Missouri Cancer Registry	passive collection since 1984
9. South Carolina Central Cancer Registry	passive collection since 1996
Beginning Collection of Benign Tumor Data	
1. North Dakota Cancer Registry	began collection 1997
2. Rhode Island Cancer Registry	began collection 1998

APPENDIX B

Codes for Primary Intracranial and Central Nervous System Tumors

Note: For this report, the designation of "primary intracranial and central nervous system tumors" includes the following primary tumors of central nervous system sites as well as tumors of the pituitary and pineal glands. All histology types within the topography codes are included in the designation of "primary intracranial and CNS tumors." The source for the topography codes is: Percy C, Van Holten V, Muir C, eds. International Classification of Diseases for Oncology, Second Edition. Geneva: World Health Organization, 1990.

Intracranial tumors

Brain:
- C71.0 Cerebrum
- C71.1 Frontal lobe
- C71.2 Temporal lobe
- C71.3 Parietal lobe
- C71.4 Occipital lobe
- C71.5 Ventricle, NOS[a]
- C71.6 Cerebellum, NOS
- C71.7 Brain stem
- C71.8 Overlapping lesion of brain
- C71.9 Brain, NOS

Meninges:
- C70.0 Cerebral meninges
- C70.9 Meninges, NOS

Cranial Nerves and other intracranial parts of the central nervous system:
- C72.2 Olfactory nerve
- C72.3 Optic nerve
- C72.4 Acoustic nerve
- C72.5 Cranial nerve, NOS
- C72.8 Overlapping lesion of brain and central nervous system
- C72.9 Nervous system, NOS

Other endrocrine glands and related structures:
- C75.1 Pituitary gland
- C75.2 Craniopharyngeal duct
- C75.3 Pineal gland

Extracranial tumors

Spinal cord:
- C72.0 Spinal cord
- C72.1 Cauda equina

Meninges:
- C70.1 Spinal meninges

[a]NOS=Not Otherwise Specified

APPENDIX C

Brain Tumor Histology Groupings

The following histology groupings were used for the primary intracranial and central nervous system tumors. The source for the histology groupings is: Central Brain Tumor Registry of the United States (CBTRUS) 1996 Statistics Report, Table 4.

WHO[a] HISTOLOGY GROUPINGS	ICD-O[b] MORPHOLOGY CODE
TUMORS OF NEUROEPITHELIAL TISSUE	
Diffuse astrocytoma (protoplasma, fibrillary)	9410, 9420
Anaplastic astrocytoma	9401, 9411
Glioblastoma	9440, 9441, 9442
Pilocytic astrocytoma	9421
Unique astrocytoma variants	9383, 9384, 9424
Oligodendroglioma	9450
Anaplastic oligodendroglioma	9451, 9460
Ependymoma/anaplastic ependymoma	9391, 9392, 9393
Ependymoma variants	9394
Mixed glioma	9382
Astrocytoma, NOS[c]	9400
Glioma malignant, NOS	9380
Choroid plexus	9390
Neuroepithelial	9381, 9423, 9430
Benign neuronal/glial, neuronal and mixed	8680/1, 8693, 9490, 9505, 9506
Malignant neruonal/glial, neuronal and mixed	8680/3, 9364, 9490, 9500
Pineal parenchymal	9360, 9361, 9362
Embryonal/primitive/medulloblastoma	8963, 9470, 9471, 9472, 9473, 9501, 9502, 9503, 9510
TUMORS OF CRANIAL AND SPINAL NERVES	
Nerve sheath, benign and malignant	9540, 9550, 9560, 9570
TUMORS OF THE MENINGES	
Meningioma	9530, 9531, 9532, 9533, 9534, 9537, 9538
Other mesenchymal, benign and malignant	8800, 8801, 8802, 8803, 8810, 8830, 8850, 8861,8900, 8910, 8990, 9133, 9150, 9240, 9480, 9481, 9536, 9539
Hemangioblastoma	9161, 9535
LYMPHOMAS AND HEMOPOIETIC NEOPLASMS	
Lymphoma	9590, 9591, 9593, 9594, 9595, 9630, 9650, 9652, 9663, 9670, 9671, 9672, 9674, 9675, 9680, 9681, 9682, 9684, 9685, 9686, 9687, 9690, 9691, 9693, 9695, 9696, 9698, 9702, 9714, 9723, 9731, 9766, 9970
GERM CELL TUMORS	
Germ cell	8020, 9060, 9064, 9070, 9071, 9080, 9084, 9085, 9100
CYSTS AND TUMOR-LIKE LESIONS	
Cysts and heterotopias	9084
TUMORS OF THE SELLAR REGION	
Pituitary	8040, 8l40, 8l46, 8260, 8270, 8271, 8280, 8281, 8290, 8300, 8330, 8323, 8333
Craniopharyngioma	9350

LOCAL EXTENSIONS FROM REGIONAL TUMORS	
Chordoma/chondrosarcoma	9370
UNCLASSIFIED TUMORS	
Hemangioma	9120, 9121, 9130, 9131
Neoplasm, benign	8000/0, 8010
Neoplasm, uncertain behavior	8000/1, 8001/1
Neoplasm, malignant	8000/3, 8001/3, 8002, 8003
All other	8720, 9580

[a]WHO = World Health Organization
[b]ICD-O = International Classification of Diseases for Oncology, Second Edition, 1990.
[c]NOS=Not Otherwise Specified

APPENDIX D

International Classification of Childhood Cancer (ICCC)

Diagnostic Group	ICD-O[a] codes	
	Morphology	Topography
I. LEUKAEMIA		
(a) Lymphoid leukaemia	9820-9827, 9850	
(b) Acute non-lymphocytic leukaemia	9840, 9841, 9861, 9864, 9866, 9867, 9891, 9894, 9910	
(c) Chronic myeloid leukaemia	9863, 9868	
(d) Other specified leukaemias	9830, 9842, 9860, 9862, 9870-9890, 9892, 9893, 9900, 9930-9941	
(e) Unspecified leukaemias	9800-9804	
II. LYMPHOMAS AND OTHER RETICULOENDOTHELIAL NEOPLASMS		
(a) Hodgkin's disease	9650-9667	
(b) Non-Hodgkin's lymphoma	9591-9595, 9670-9686, 9690-9714, 9723	
(c) Burkitt's lymphoma	9687	
(d) Miscellaneous lymphoreticular neoplasms	9720, 9731-9764	
(e) Unspecified lymphomas	9590	
III. CENTRAL NERVOUS SYSTEM (CNS) AND MISCELLANEOUS INTRACRANIAL AND INTRASPINAL NEOPLASMS[b]		
(a) Ependymoma[c]	9383, 9390-9394	
(b) Astrocytoma	9380	C72.3
	9381, 9400-9441	
(c) Primitive neuroectodermal tumours	9470-9473	
(d) Other gliomas[d]	9380	C70.0-C72.2, C72.4-C72.9
	9382, 9384	
	9442-9460, 9481	

Diagnostic Group	ICD-O[a] codes	
	Morphology	Topogragphy
III. CNS AND MISCELLANEOUS INTRACRANIAL AND INTRASPINAL NEOPLASMS,[b] Continued		
(e) Miscellaneous intracranial and intraspinal neoplasms[c]	8270-8281, 8300, 9350-9362, 9480, 9505, 9530-9539	
(f) Unspecified intracranial and intraspinal neoplasms[c]	8000-8004	C70.0-C72.9, C75.1-C75.3
IV. SYMPATHETIC NERVOUS SYSTEM TUMOURS		
(a) Neuroblastoma and ganglioneuroblastoma	9490, 9500	
(b) Other sympathetic nervous system tumours	8680, 8693-8710, 9501-9504, 9520-9523	

[a]International Classification of Diseases for Oncology, Second Edition. World Health Organization, 1990.
[b]Group III excludes all lymphomas. See group II.
[c]Behaviour codes /0 and /1 are included.
[d]Behaviour code /1 is included.